Supravalvular Congenital Mitral Stenosis

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Congenital supravalvular mitral stenosis is a rare malformation characterized by the presence of a shelf-like fibrous membrane, with 1 or 2 small orifices, covering and obstructing the mitral valve. The membrane is positioned closely to the mitral valve (and sometimes it is attached to it); therefore, a preoperative diagnosis is inevitably difficult, even with the use of biplane echocardiography. Two patients with supravalvular mitral stenosis aged 3 years and 3 months are described. In 1 patient, a preoperative diagnosis was made, and both successfully underwent correction.

Supravalvular congenital mitral stenosis is a rare malformation, described by Fisher[1] in 1902, and is anatomically characterized by the presence of a stenosing ring in the form of a shelf-like membrane above the mitral valve. This membrane has 1 or 2 small orifices, which obstruct the left atrial outflow. The mitral valve may be normal or deformed. This condition has to be distinguished from abnormal partition of the left atrium (*cor triatriatum*), in which a membrane divides this cavity into 2 chambers: a dorsal chamber, that receives the pulmonary veins and a ventral chamber that gives rise to the left atrial appendage.

We report here 2 cases of supravalvular mitral stenosis treated surgically. In one, the diagnosis was made prior to surgery.

Case report

Case 1 - A 3-year-old male was born of a cesarean delivery with normal development. He experienced progressive dyspnea on effort 1 year previously, and more recently, he experienced nocturnal dyspnea. On physical examination, he was found to be in good overall condition and weighed 15kg. His pulse was palpable in the 4 limbs with systemic blood pressure of 90x60mm Hg. A sharp precordial bulging was observed. Cardiac rhythm was regular at 80bpm. A short diastolic thrill with presystolic accentuation was heard in the mitral area. The pulmonic component of the second sound and first mitral sound were accentuated. No rates were available on pulmonary auscultation and other physical findings were within normal limits. The electrocardiogram revealed sinus rhythm and evidence of right ventricle hypertrophy. The chest X-ray showed intensive pulmonary plethora and evidences of left atrium and rightventricle enlargement. The child underwent cardiac catheterization that showed high pressure in the right ventricle (74x5mmHg) and pulmonary artery (78x10mmHg). Left cineventriculography did not show abnormalities. On July 4, 1989, the child underwent surgery with a diagnosis of congenital mitral stenosis. Open-heart surgery was performed through a midline sternal splitting incision with total cardiopulmonary bypass. Myocardial protection was performed with crystalloid cardioplegia and topical heart hypothermia. The enlarged left atrium was longitudinally opened, enabling fibrous membrane identification, located immediately above the mitral valve, with a central opening of 4mm in diameter. This membrane was carefully dissected. The mitral valve was normal. The left atrium was restored and the procedure was ended as usual. The postoperative period was uneventful. Almost 12 years after the surgery, the patient remains asymptomatic, and both physical examination and echocardiogram are normal.

Case 2 - A 3-month-old female, the product of a normal 38-week delivery, was admitted with frank congestive heart failure. The mother reported that the infant experienced tiredness since birth. On physical examination, the infant was found to have dyspnea, dehydration, and paleness. She weighted 4.500g. Her respiratory rate frequency was 44/min. Her cardiac rhythm was constant at 180bpm. Her pulse was palpable in the 4 limbs-with systemic blood pressure 70x40mmHg. No murmurs could be heard. The second heart sound had a loud pulmonary component. The liver was

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palpable 4 cm below the right costal margin. The chest X-ray showed a cardiac area within normal limits and accentuated pulmonary plethora. The electrocardiogram revealed sinus rhythm and was suggestive of both left atrium and right ventricle hypertrophy. The echocardiogram showed great left ventricle enlargement and an image suggestive of a supravalvular mitral membrane (fig. 1). Based on this diagnosis, the child underwent surgery on October 23, 2000. Through a median sternotomy incision, cardiopulmonary bypass was installed and myocardial protection was performed with crystalloid cardioplegia and topical hypothermia of the heart. The left atrium was very enlarged. A left atriotomy enabled identification of a membrane right above the mitral valve with 2 small openings that allowed blood to flow from the atrium to the left ventricle (fig. 2). This membrane was excised enabling access to a normal mitral valve (fig. 3). The left atrium was restored, and the surgery ended as usual. Follow-up was normal, and after 6 months, the child was asymptomatic with normal physical development.

Discussion

Congenital supravalvular mitral stenosis is a rare malformation frequently associated with other cardiac defects,

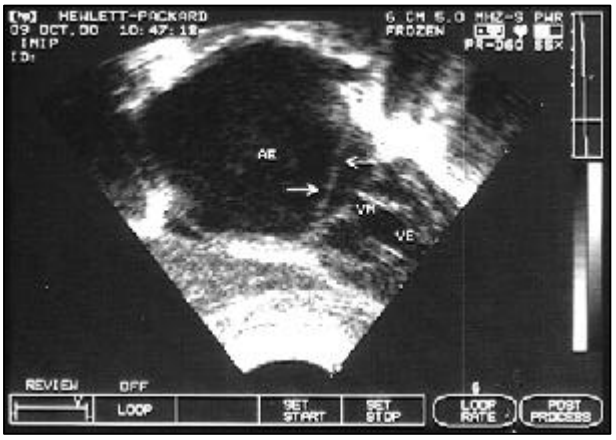

Fig. 1 - Case 2 echocardiogram in which we can clearly observe supravalvular mitral membrane.

Fig. 2 - Surgical aspect of mitral supravalvar membrane.

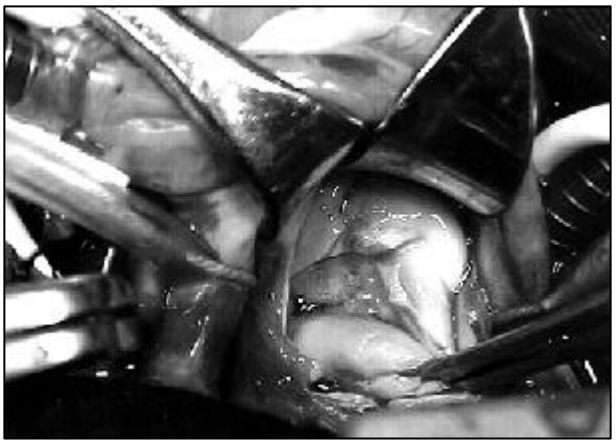

Fig. 3 - Surgical aspect of mitral valve after membrane resection.

such as interventricular communication, coarctation of the aorta, subaortic stenosis, parachute mitral valve, left superior vena cava draining in the tectum of the left atrium and *cor triatriatum* [2-8]. Even more rare is its isolated occurrence, such as in the cases reported here, first described by Chung et al [9] in 1974.

The physiologic consequence of a supravalvular mitral membrane is similar to that of other obstructive abnormalities that can occur in the left atrium (pulmonary vein stenosis, *cor triatriatum*, mitral valvular stenosis), and it includes increases in venocapillary pressure as well as pulmonary arterial pressure. Children who have this disease present with precocious congestive heart failure. Clinical signs as well as electrocardiography, radiology, and hemodynamic alterations suggest mitral valve stenosis. Angiographic study occasionally shows the membrane [6]. Preoperative identification of the lesion is easier with biplane echocardiography, which is the best method to identify the anatomy of the obstructions of the left heart [10]. However, technical limitations exist to visualization of the membrane that is usually smaller than 1 mm and is either very close or attached to the mitral valve [7]. A preoperative diagnosis was done in less than half of the operated cases [7]. In one of our patients, we could clearly identify the supravalvular membrane through echocardiography.

The first surgical correction of congenital supravalvular mitral stenosis was described by Lynch et al [2], in 1962 in a study of 14 patients operated on at the Hospital for Sick Children [7] in London. The study showed that surgical correction of this anomaly, with or without associated malformations, can be successfully performed in most cases, leading to excellent late clinical results. Both cases presented here confirm these data, indicating that surgery must be performed without delay in all children with evidence of left atrial outflow obstruction. In contrast with other forms of congenital mitral stenosis that can be corrected by percutaneous valvuloplasty, surgery seems to be the only possible treatment for supravalvular stenosis.

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